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CASE REPORT

Cervical myelopathy caused by soft-tissue mass in diffuse idiopathic skeletal hyperostosis

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Abstract A rare case of cervical spinal cord compression in diffuse idiopathic skeletal hyperostosis (DISH or Forestier's Disease) caused by a craniocervical mass of soft-tissue is reported. The objective is to describe an uncommon mechanism of spinal cord compression in DISH. Three weeks after a cardiac infarction a 69-year-old man slowly developed spastic tetraparesis. Magnetic resonance tomography showed a craniocervical tumor compressing the spinal cord and a massive DISH of the cervical spine. An extended mass of yellowish amorphous material was removed from between the dura, the posterior odontoid process and the posterior aspect of vertebral body C2 reaching to the upper part of C3.The histologic appearance indicated connective tissue and cell-degenerated cartilaginous tissue. There was no inflammatory component and no evidence of neoplasia. No ossification of the posterior longitudinal ligament (OPLL) was found. After removal and craniocervical stabilization the patient's neurologic function improved remarkably. The increase of mechanical stress on the atlantoaxial segment and enhanced proliferation reaction of the connective tissue in DISH are suggested as the underlying pathomechanisms in the formation of this soft-tissue mass.

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Introduction

Diffuse idiopathic skeletal hyperostosis (DISH), also termed Forestier's disease, occures in about 12% of middle-age and elderly men [24]. Anterolateral ossification of the anterior longitudinal ligament leads to ankylosing hyperostosis of certain continuous vertebral bodies [19, 26]. Besides the reduced mobility of the affected parts of the spine, the most common symptoms of cervical DISH are compression of the trachea and esophagus [3, 12, 23]. Neurological symptoms such as paresthesias or motor disturbancies, are found in only 4% of the patients [27]. The hyperostosis of spine can be associated with impairment of spinal cord, nerve root compression [1, 4, 5, 14, 15, 29] or vertebral artery insufficiency [22]. Reports of cervical myelopathy in DISH are rare. A few cases of cervical spinal cord compression due to atlantoaxial subluxation [4, 16], odontoid fracture [7], pseudotumor [9], ligamentous hypertrophy [18] and basilar impression [17] have been reported.

Case report

A 69-year-old man presented with gradual disturbance of gait due to spastic tetraparesis over a 3-month period. Symptoms had begun 3 weeks after surgery for cardiac infarction consisting of balloon angioplasty of the heart vessels and stent procedure, which was performed without general anesthesia. Initially the patient noticed decreased

sensation in the finger tipps of both hands. Later on, there was a rapid progredient weakness and stiffness of both legs. Finally, when being hospitalized in our clinic 12 weeks after beginning of the symptoms, he could only walk a few steps unassisted. He reported loss of sensation in parts of the left leg and the left testicle. He felt electrifying pain in his left leg on turning his head to the left.

Examination showed a positive Lhermitte sign and spastic tetraparesis accentuated to the left arm and leg. The patient's reflexes were brisk. Babinki's sign was present on both sides. Touch and pain sensation was decreased in all fingers, in anterior parts of the left leg and in the left testicle. Sensation was not altered in the patients' trunk. There were no signs of autonome dysreflexia. The patient reported rapid detoriation in the last 2 weeks.

The patient did not show clinical, radiological, or serological evidence of rheumatoid arthritis or of ankylosing spondylitis (Bechterew's disease). Radiological signs of osteoarthritis were seen in both shoulders without clinical symptoms. Radiographs of both hips, hands and shoulders showed no calcification, so there were no hints for calcium pyrophosphate deposits.

Computed tomography of the cervical spine showed massive hyperostosis between C2 and C3 and from C4 to C7 (Fig. 5).

A sagittal T1-weighted MRI showed a large, smooth, intermediate-signal mass reaching from behind the odontoid process to the upper part of C3 compressing the high cervical spinal cord (Fig. 1). The axial T2-weighted image shows this mass stretching across the anterior half of the spinal canal (Fig. 2).

The T2-weighted sagittal image shows that the mass consisted of materials varying in signal intensity (Fig. 3).

Because of impending paraplegia surgery of cervical spine was performed as an emergency procedure accepting an elevated risk since the heart attack was only 16 weeks ago. The extradural mass was removed via lateral approach. Compared to the most commonly used transoral procedure the lateral approach provides a shorter operative route, avoids complications by contaminated nasophyrynx and allows immediate occipitocervical fusion [11]. Approximately a third of the posterior C1 arch had to be removed (Fig. 4). The postoperative sagittal computed tomography shows the whole extent of the hyperostosis of cervical spine (Fig. 5).

A glassy yellowish mass of soft material extruded on incision of the posterior longitudinal ligament. The histological appearance (Fig. 6) indicated connective tissue and cell-degenerated cartilaginous tissue. There was no focal proliferation of small vessels, no inflammatory component, no evidence of neoplasia and no calcium pyrophosphate dihydrate crystals. No OPLL was found. To stabilise the atlantoaxial segment, an occipitocervical fusion using a titanium frame was performed (Fig. 7).The patient's neurological function improved remarkably.

Three weeks after the decompression the patient could walk 800 m with two walking sticks. The loss of pain and touch sensation of the left arm and leg persisted.

Fig. 1 The preoperative T1-weighted sagittal magnetic resonance spin-echo image shows a large smooth, intermediate signal intensity mass reaching from behind the odontoid process down to the upper limit of C3

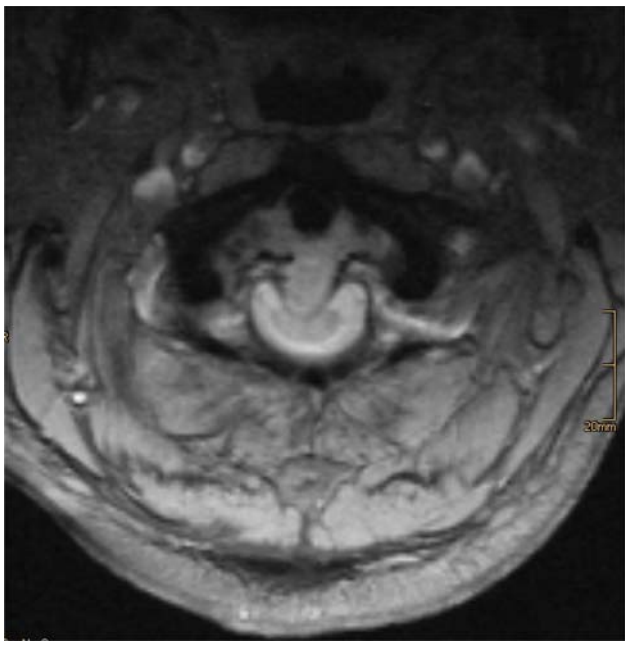

Fig. 2 Preoperative T2-weighted axial magnetic resonance spin-echo showing an inhomogenous smooth mass stretching over about half of the spinal canal and compressing the cervical spinal cord

Fig. 3 The preoperative T2-weighted sagittal magnetic resonance image shows that the mass consisted of materials varying in signal intensity

Fig. 4 Postoperative axial computed tomography showing that about a third of the posterior arch of C1 was removed

Discussion

DISH is commonly associated with ossification of the anterior longitudinal ligament [2, 19, 28] and is often related to ossification of the posterior longitudinal ligament (OPLL), a multifactorial disease in which complex genetic and environmental factors interact [8]. DISH of the cervical spine affects the lower region between C4 and C7 much more than the upper region between C1 to C3. A study of 68 patients with cervical DISH showed only 10.2% manifestation at C1 and segmental motion of atlantoaxial joint was preserved in 90% of the patients [20, 21].

Cervical DISH is therefore associated with reduced mobility of the lower cervical spine thus leading to enhanced mechanical stress of the still-mobile upper parts. This could explain cases of odontoid fractures [7] or of atlatoaxial subluxation [4, 16] in DISH.

Enhanced strain to the still-mobile occipitoatlantoaxial segment could lead to partial tears of the ligaments followed by chronic repair processes thus forming hypertrophic masses of soft tissue [6, 18]. As reported,

Fig. 5 Postoperative sagittal computed tomography showing the whole extent of hyperostosis bridging C3–C7

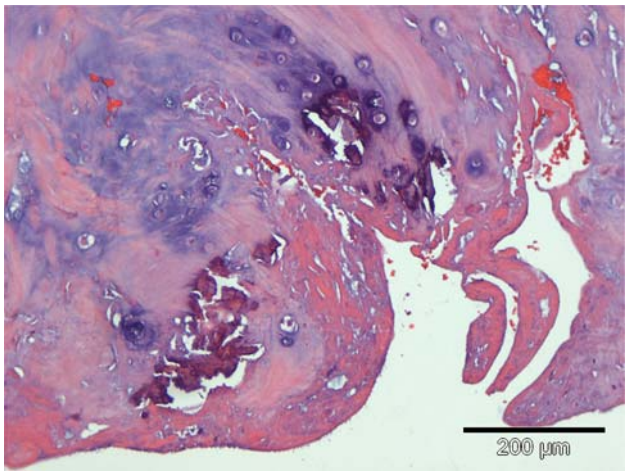

Fig. 6 Histologic appearance of the removed tissue, hematoxilin-eosin staining

Fig. 7 Postoperative lateral radiograph of the cervical spine showing the titanium frame used for stabilization

formation of soft tissue and fibrocartilaginous metaplasia is caused by localized trauma or chronic mechanical stress [15]. Enhanced proliferative reaction of the connective tissue, an early change occurring in DISH prior to ossification, may have contributed to the formation of the large tissue mass found in our patient [24]. Periodontoid inflammatory soft-tissue can reportedly be caused by chronic inflammation [6], for example in rheumatoid arthritis (rheumatoid pannus).

Our patient did not show features of rheumatoid arthritis. The histological appearance indicated connective tissue without ossification and cell-degenerated cartilaginous tissue, so there were no inflammatory components in this case and no hints for OPLL. Histologic findings in our case are very similar to a case of retro-odontoid pseudotumor described by Jun By et al. [9]. Studies have shown that periodontoid soft tissue masses caused by rheumatoid arthritis or by trauma can be reduced by posterior fixation alone [10, 13, 25, 30]. In our patient additional removal of the tumorous structure was performed because of severe, progressing myelopathy and of uncertainty about the diagnosis.

Magnetic resonance tomography had suggested that the inhomogenous process were a non-neoplastic and fibrous structure, but an inflammatory pannus or a soft tissue sarcoma could not definitively be excluded.

Conclusions

This case shows an uncommon mechanismn of cervical myelopathy in DISH not due to ossification of the posterior longitudinal ligament (OPLL) but to a large mass of soft-tissue. Only a few comparable cases of ligamentous hypertrophic changes with myelopathy [18] and one case of a retro-odontoid pseudotumor in DISH [9] are described.

We suggest that transfer of mechanical stress to the atlantoaxial segment was one of the underlying pathomechanisms for the formation of this soft-tissue mass.

Hypertrophy of soft tissue as a precursor of ossification may be a common feature in the often coexisting diseases DISH and OPLL.

Conflict of interest statement None of the authors has any potential conflict of interest.

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